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Managing and coping with Hemophilia

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Hemophilia is an inherited disorder where the blood does not clot normally as it should. This disorder is because of a defect in one of the clotting factor genes on the 'X' chromosome.

Every year 17th April is recognized as World Hemophilia Day to increase awareness about hemophilia and other inherited bleeding disorders, as well as to improve access to care and treatment. Hemophilia is an inherited disorder where the blood does not clot normally as it should. This disorder is because of a defect in one of the clotting factor genes on the 'X' chromosome.

What is hemophilia?

Hemophilia is an inherited bleeding problem, where those affected do not bleed faster than normal, but they can bleed for a longer time. Their blood does not have enough clotting factor which is a protein in blood that controls bleeding.

Types of hemophilia

- Hemophilia A is the most common type of hemophilia. This means the person does not have enough clotting factor VIII (factor eight).
- Hemophilia B is less common. Here the person does not have enough factor IX (factor nine). In both hemophilia A and B patient's bleed for a longer time than normal.

Degrees of hemophilia

- Severe hemophilia: sufferers usually bleed frequently (one to two times a week) into their muscles or joints. Bleeding is often spontaneous, which means it happens for no obvious reason.
- Moderate hemophilia: patients bleed less frequently (about once a month), and may bleed for a long time after surgery, a bad injury, or dental work. They will rarely experience spontaneous bleeding.

- People with mild hemophilia usually bleed as a result of surgery or major injury. They do not bleed often and some may never show symptoms.

Symptoms and diagnosis

The signs of hemophilia A and B are the same:

- Big bruises
- Bleeding into muscles and joints
- Spontaneous bleeding (sudden bleeding inside the body for no clear reason)
- Prolonged bleeding after getting a cut, removing a tooth, or having surgery
- Bleeding for a long time after an accident, especially after an injury to the head

Bleeding into a joint or muscle causes:

- Swelling
- Pain and stiffness
- Difficulty using a joint or muscle

Where does bleeding occur?

Excessive bleeding and bruises are the main symptoms of Hemophilia. People with hemophilia commonly have internal bleeding, into muscles or joints. The most common muscle bleeds occur in the muscles of the upper arm and forearm, the iliopsoas muscle (the front of the groin area), the thigh, and the calf. The joints that are most often affected are the knee, ankle, and elbow.

"Repeated bleeding into the same joint can lead to pain, permanent damage and arthritis, so walking is often affected. However, the joints of the hands are not usually affected in hemophilia," said Dr. Satish Kumar A, Consultant – Oncology, Columbia Asia Referral Hospital Yeshwanthpur.

How is hemophilia diagnosed?

Hemophilia is diagnosed by taking a blood sample and measuring the level of factor activity in the blood. Hemophilia A is diagnosed by testing the level of factor VIII activity. Hemophilia B is diagnosed by measuring the level of factor IX activity.

If the mother is a known carrier of hemophilia, testing can be done before a baby is born. Prenatal diagnosis can be done at 9 to 11 weeks by chorionic villus sampling (CVS) or by fetal blood sampling at a later stage (18 or more weeks).

Treatment

The missing clotting factor is injected into the bloodstream using a needle. Bleeding stops when enough clotting factor reaches the spot that is bleeding.

Bleeding should be treated as quickly as possible. Quick treatment will help reduce pain and damage to the joints, muscles, and organs. If bleeding is treated quickly, less factor is needed to stop the bleeding.

With an adequate quantity of treatment products and proper care, people with hemophilia can live perfectly healthy lives.

- Dr. Satish Kumar A, Consultant – Oncology, Columbia Asia Referral Hospital Yeshwanthpur